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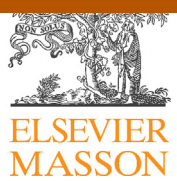

LETTER / *Muskuloskeletal imaging*

Dipygus: Computed tomography findings and management

Keywords Dipygus; Supernumerary feet; CT; Burkina Faso

Dear Editor,

Complete dipygus is a very rare congenital malformation involving complete lumbosacral duplication with two pelves and four lower limbs [1]. Several incomplete forms have been reported. Pygomelia is an incomplete form in which two of the lower limbs fusion (symmelia) form a single supernumerary middle limb inserting into the pelvis [2—5]. Pygomelia has often been described as caudal or lumbosacral duplication with a supernumerary limb [6—11]. Other reports have described even less complete forms with no accessory limbs, the only evidence of dipygus being a duplication of external genitalia and the anal orifice [6—9]. Due to the rareness of this condition, very few cases have been described in the literature [2—13], particularly in Africa where only three cases have been reported up to now [1—3]. Furthermore, the use of the terms dipygus, pygomelia and caudal duplication is not always clear in African literature and there has been some confusion [2—13]. We report here the imaging findings of a case of incomplete dipygus using multidetector row computed tomography (MDCT), and discuss the issues related to its management.

Case report

A 33-day-old female infant was referred to the medical imaging department for evaluation of a congenital malformation. The infant was born by normal vaginal delivery after full-term pregnancy. The pregnancy had not been supervised and the mother received no antenatal visits, obstetrical ultrasound examinations, or serological tests. There was no mention of dystocia or other complications during labor. The infant was the first child of a young couple. There was no family history of malformation or consanguinity, or record of exposure to medicinal products or other teratogens during pregnancy.

Physical examination of the infant demonstrated a good nutritional status and a normal appearance on the anterior view (Fig. 1). The child weighed 3760 g and measured 48 cm. The patellar and Achilles reflexes of the lower limbs, as well as the abdominal and cremasteric reflexes, and the anal wink were normal. Examination revealed the presence of a voluminous mass (Fig. 1) of approximately 21 cm in diameter in the lumbosacral region into which were implanted a supernumerary left foot and right toe separated by a groove (Fig. 1). The two supernumerary lower limbs were found to be sensitive and could be mobilized by the infant; however they were moved discordantly one from another and from the other limbs. Gynecological examination was performed with normal results.

X-ray examinations of the lumbosacral spine did not provide any significant information allowing to describe the etiology of the malformation. The findings of transfontanellar cranial and abdominal ultrasound examinations were normal.

MDCT of the abdomen and pelvis was performed before and after intravenous administration of iodinated contrast medium. Multiplanar reconstructions revealed that although solid visceral organs were normal, the infant

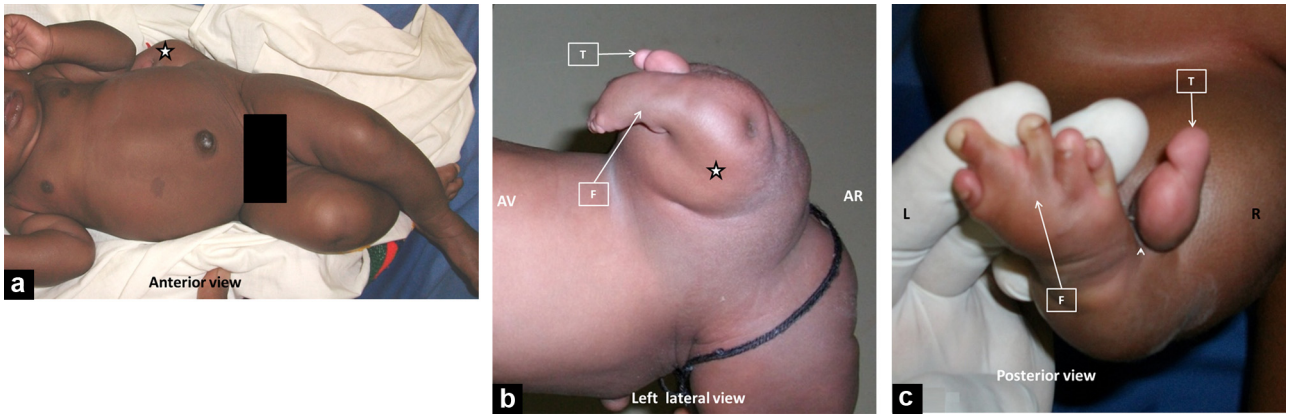

Figure 1. Photographs of a 33-day-old infant presenting an incomplete form of dipygus. Normal anterior morphological appearance (a); lumbosacral mass (a, b, c, star) to which is attached a left foot (b, F) and a right toe (c, T) separated by a groove (c, arrowhead). F: left foot; T: right toe; a: anterior view; b: left lateral view; c: posterior view.

showed spinal dysraphism at the L3, L4 and L5 levels with tethered cord attached at a low point and spreading into the sacral region where it was enclosed in a voluminous subcutaneous mass of fatty density (Fig. 2). The mass also contained a duplicated, inversely oriented, non-adherent hemi-pelvis adjacent to the dysraphism. The extra hemi-pelvis was innervated by nerve roots from the conus medullaris (Fig. 2). The whole left foot which included five toes was articulated into the duplicated pelvis by two ossification centers within the acetabulum. One of these could be the talus and the other the calcaneus (Fig. 3). The right toe was articulated into the developing ilium via a ''neo joint'' with an ossification center that could be the talus (Fig. 3).

The hypothesis of conjoined dipygus twins was suspected and it was decided that surgery was indicated following a multidisciplinary staff meeting (radiologist, pediatrician, orthopedic surgeon and neurosurgeon). Nevertheless, despite prior psychological preparation, the child's parents refused surgical management.

Discussion

No similar case was found in the literature. It should be noted that various different terms including dipygus, pygomelia, monstrosities, lumbosacral duplication with an extra lower limb and caudal duplication, are used to denote this kind of malformation [2—13]. Whatever the term used, it remains a very rare congenital malformation [14]. Its true prevalence remains unknown, although the prevalence of congenital anomalies of the lower limbs can be estimated to 1/100,000 births [5].

We consider that the case described here is an incomplete form of dipygus. Indeed, in its complete form, dipygus results in an infant with a single head and trunk that

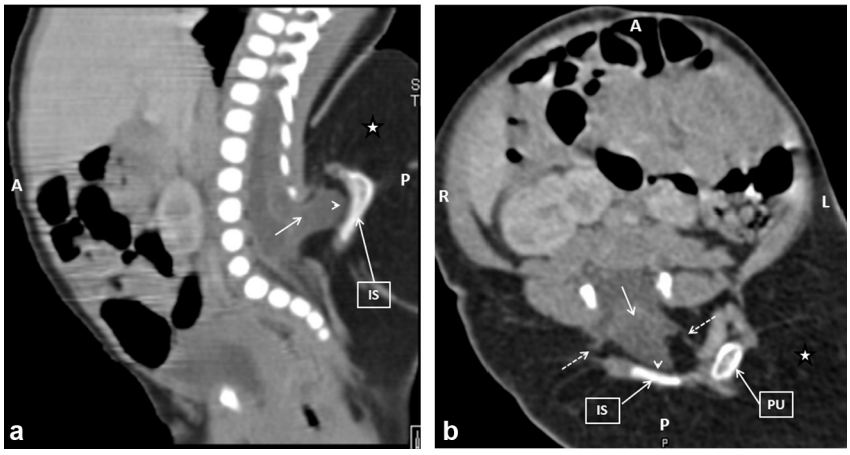

Figure 2. Abdominopelvic MDCT examination after injection of iodinated contrast material. Spinal dysraphism (a, b, full arrow) with tethered cord with a low attachment point on the malformation (a, b, arrowhead) showing nerve roots (a, b, dashed arrow) and lumbosacral malformation (b, c, star). IS: ischium; PU: pubis; a: sagittal reformation in MPR mode; b: axial slice in MPR mode.

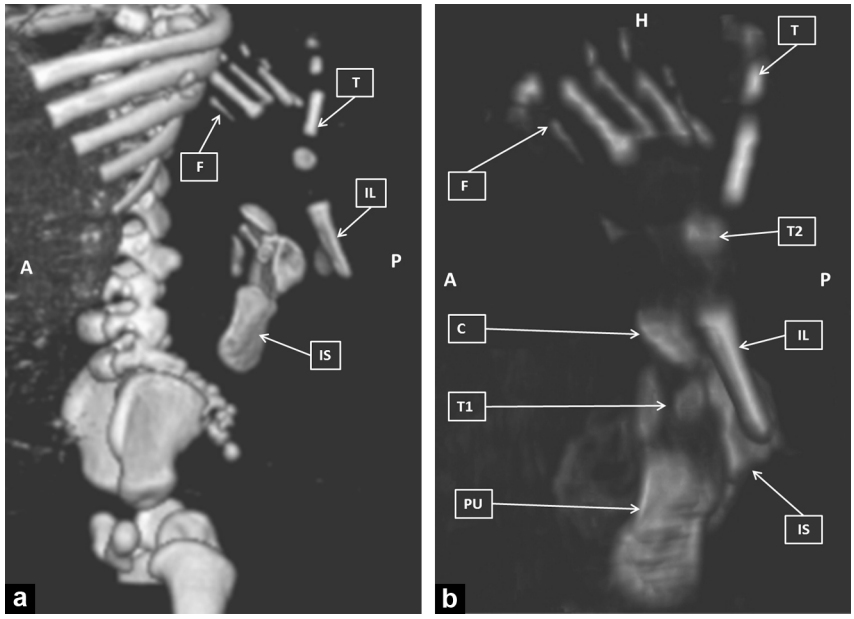

Figure 3. Three-dimensional (3D) MDCT images. a: 3D MDCT image; b: 3D CT image with suppression of autosite. F: left foot; T: right toe; IL: ilium; IS: ischium; PU: pubis; T1: left talus; T2: right talus; C: left calcaneus.

becomes duplicated from the lumbosacral level down, with two pelves, two pairs of buttocks and four legs [1]. In our case, the incomplete duplication was materialized by an additional hemi-pelvis, a significant lumbosacral mass with two dimples corresponding to the extra buttocks and two supernumerary lower limbs represented here by just the feet. In other reports of incomplete dipygus, the two lower limbs fusion axially by symmelia to give a single accessory limb that inserts into the pelvis, this form is generally called three-limbed pygomelia [2—4]. Other even less complete forms of dipygus have also been described, in which the only visible evidences of dipygus are duplicated urogenital organs and two anuses [1]. Such cases of duplication are forms of incomplete twinning where the parasitic twin is attached to the caudal region of the host twin (also called independent twin or autosite). The case described in this paper is that of asymmetrical Siamese sisters, the lumbosacral mass being the underdeveloped parasitic twin.

The frequency of conjoined twins is of approximately 1 case for 30,000 to 100,000 births. In 70% of cases, conjoined twins are females [1]. Imaging is crucial for an appropriate assessment of dipygus. Indeed, standard X-ray examination is the main imaging modality used to assess bone formation or segmentation anomalies [4,5]. However, as for our patient, its contribution is limited when it comes to understanding the etiology of complex malformations. Due to the unavailability of MR imaging in our hospital, MDCT was therefore performed despite the exposure to radiation. Multiplanar reformations allowed a comprehensive evaluation of the malformation, identified tethered cord with a low attachment point and advised suitable surgical management. Unfortunately, surgery was not performed due to the parents' refusal to give consent, even after appropriate psychological preparation.

Conclusion

In conclusion, dipygus is a very rare congenital malformation in humans. To our knowledge, this is the first description of a case of articulated lumbosacral duplication with two supernumerary feet. The diagnostic was made following anatomical assessment of the malformation based on multiplanar reconstructions of MDCT data and discussions with various teams of specialists.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

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